The Open Journal of Occupational Therapy

Volume 9
Issue 2 *Spring 2021*

Article 5

April 2021

Home Use of an Upper Extremity Exoskeleton in Children with SMA: A Pilot Study

Tracy M. Shank
Nemours/Alfred I. duPont Hospital for Children – USA, trashank29@gmail.com

Tariq Rahman
Nemours/Alfred I. duPont Hospital for Children – USA, trahman@nemours.org

Recommended Citation

Shank, T. M., & Rahman, T. (2021). Home Use of an Upper Extremity Exoskeleton in Children with SMA: A Pilot Study. *The Open Journal of Occupational Therapy, 9*(2), 1-14. https://doi.org/10.15453/2168-6408.1762

Home Use of an Upper Extremity Exoskeleton in Children with SMA: A Pilot Study

Abstract

Background: People with spinal muscular atrophy (SMA) often have arm weakness resulting in restricted independence and challenges with activities of daily living. An upper extremity (UE) orthosis, the Wilmington Robotic Exoskeleton (WREX), which augments arm movement by providing gravity assistance, was provided to a small cohort of subjects for 1 year. Resulting changes in the subjects' performance were assessed.

Method: Five subjects with SMA were asked to use the WREX system for 1 year. Data were collected at baseline and at 6-month intervals. Evaluation tools used were UE range of motion (ROM), the Box and Block Test, the Canadian Occupational Performance Measure (COPM), and the reachable surface area (RSA) using a Microsoft Kinect Sensor.

Results: There were no significant changes in UE ROM without the device over time and no significant changes in dexterity after long-term use of the WREX. There were clinically meaningful changes in active ROM while wearing the device compared to without it and clinically meaningful changes in performance and satisfaction while wearing the device. The RSA software did not yield usable results for this population.

Conclusion: Wearing bilateral WREX devices resulted in immediate improvements in ROM and function; however, the subjects experienced several barriers, which prevented consistent long-term use.

Keywords

spinal muscular atrophy, exoskeleton, orthosis, assessment

Cover Page Footnote

Financial support was provided by CureSMA, Grant # RAH1617

Credentials Display

Tracy M. Shank, MS, OTR/L, CHT

Tariq Rahman, PhD

DOI: 10.15453/2168-6408.1762

Many neuromuscular conditions negatively impact children's activities of daily living (ADLs) and leisure tasks. This progressive decline in performance can lead to feelings of frustration and dependency and, if left untreated, may cause patterns of disuse that could result in pain and contractures. Spinal muscular atrophy (SMA) is one of these neuromuscular conditions. It is an autosomal recessive motor neuron disease that occurs in one in every 10,000 births (SMA, 2019). It is characterized by degeneration of spinal motor neurons resulting in progressive muscle wasting and loss of movement. The severity of symptoms varies by type with Type 1 being the most severe and Type 4 being the least severe. Individuals with Type 1 SMA generally present with onset in the first 6 months of life, and median survival is only 1 year. Babies are typically normal developing at birth and have normal cognition; however, as spinal motor nerves quickly degenerate, severe weakness and respiratory distress interrupt development and cause death. Type 2 SMA presents by 18 months of age. Children with this form typically will learn to sit but never walk. Type 2 SMA represents about 20% of cases, and the median survival rate is upward of 25 years. The lower extremities usually are affected more than the upper extremities, and cognition is normal. Children in this group frequently present with impaired swallowing and ventilatory insufficiency. Children with Type 2 SMA will develop scoliosis, which will impair function further as they get older. Type 3 SMA generally presents in children over 18 months of age, and these children may learn to stand or even walk in childhood but become wheelchair-bound as the disease progresses. This subtype makes up 30% of cases. People with SMA Type 3 have a normal life span. Type 4 SMA represents only 5% of cases, and it is the mildest form of the disease with onset after 30 years of age and normal life span (Arnold et al., 2015). The subjects recruited for this study were all diagnosed with SMA Type 2.

Treatment for children with SMA typically includes physical and occupational therapy; durable medical equipment, such as power wheelchairs, bath seats, activity chairs, Hoyer lifts, and braces; respiratory therapies and close following by a pulmonologist; orthopedic care to address contractures and scoliosis in later childhood and adolescence; and genetics, nutrition, and neurology (SMA, 2019).

Literature Review

Children with SMA Type 2, as described earlier, demonstrate progressive muscle weakness over time. Functionally speaking, a child may be able to swing their arm using momentum to achieve shoulder flexion, but they will not have the strength to support the arm in space to complete a task. In addition, if the child is holding an object, their endurance is further reduced. Activities done at table level, such as playing with blocks, can quickly cause fatigue resulting in a loss of function. One way for a child with SMA to improve access to their environment is through the use of technology, such as an arm exoskeleton. This technology is discussed below.

Upper Extremity Devices

There are a few upper extremity (UE) exoskeleton systems that have been designed to augment movement, and some have been commercialized (Van der Heide et al., 2014) (see Table 1). The more successful systems include the ARMON, which is a wheelchair-mounted exoskeleton that allows the arm to move against gravity. The ARMON (Armon Products, 1999) is powered by adjustable springs and is used for people with neuromuscular conditions (Herder et al., 2006). The Dynamic Arm Support (Assistive Innovations, n.d.) is a wheelchair-mounted, spring-loaded orthosis for people with arm weakness. The SaeboMAS (Saebo Inc., n.d.) is also a gravity-compensated arm support that is commercialized. There is little information on the long-term evaluation of these devices. The Wilmington Robotic EXoskeleton (WREX) (JAECO Orthopedic, n.d.) is the most affordable option and has seen commercial success. This study examines long-term evaluation of the WREX for subjects with SMA.

Table 1
Some Current Commercial Upper Extremity (UE) Devices Available

	ARMON	SaeboMAS	Dynamic Arm Support	WREX
Power	Spring and electric	Spring	WC battery	Rubber band
Attachment	WC, table, activity chair	Table or floor	WC or table	WC, table, activity chair, TLSO in very young children
Population	Neuromuscular, stroke	Neuromuscular, stroke, cerebral palsy	Stroke, cerebral palsy	Neuromuscular, stroke, arthrogryposis
Cost	~$5,000	$5,800	$28,000	$2,200

Note. WREX = Wilmington Robotic EXoskeleton; WC = wheelchair; TLSO = thoracolumbosacral orthosis; MAS = mobile arm support.

The WREX is a body-powered, four degrees of freedom orthosis that provides gravity eliminated movement by using rubber bands to create a statically balanced mechanism negating the weight of the arm (Haumont et al., 2011; Rahman et al., 2006). This system allows someone with muscle strength of only 2 out of 5 to move their arm in space against gravity to perform functional tasks. The orthosis consists of a set of linkages and joints that sits alongside the child's arm and can be attached to a wheelchair or an independent mount. For young ambulatory children, the WREX has been attached to a thoracolumbosacral orthosis (TLSO). The WREX (JAECO Orthopedic, n.d.) orthosis can be purchased in adult and pediatric sizes and can be used either unilaterally or bilaterally. The pediatric and adult versions both have adjustable features, including telescoping arm lengths and changeable gravity compensation by changing the number of bands. These adjustments allow it to be customized for each patient (Rahman et al., 2007) (see Figure 1).

Figure 1
JAECO WREX in Use with Patient

Several studies have looked at functional change with use of the WREX in populations of children with mixed neuromuscular conditions (Gunn et al., 2016; Rahman et al., 2006; Shank et al., 2017). These studies have described positive change in immediate function on fine motor tasks with the Jebsen-Taylor Hand Function Test and with self-identified meaningful activities using the Canadian Occupational Performance Measure (COPM) when wearing the WREX either bilaterally or unilaterally when compared with not wearing a device. While each of these studies measured functional change in performance of everyday activities, none of them have followed patients over time to look at changes in progress or barriers. This pilot study, funded by a grant from the Cure SMA Foundation, was performed to help define the course of future larger-scale studies to better define appropriate patient populations, identify successes and barriers to UE orthotic procurement and use over time, and see the effect (if any) on disease progression. Our objectives were as follows:

1. To determine if regular use of an UE exoskeleton changes the quality of movement and/or gross dexterity of people with SMA over the course of 1 year.
2. To determine if use of an UE exoskeleton achieves clinically meaningful changes in function and satisfaction.
3. To determine if regular use of an UE exoskeleton influences long-term changes in ROM at the shoulder or elbow, which are common because of disuse.

Method

Subjects

Five subjects (four girls and one boy) were recruited from the Midwest, Northeast, and Mid-Atlantic sections of the US. The subjects were recruited by placing flyers at the Muscular Dystrophy Association muscle clinic at the Nemours/Alfred I. duPont Hospital for Children in Wilmington, DE, and at the pediatric neuromuscular clinic at Columbia University in New York City. The study was approved by the Nemours Institutional Review Board. All subjects appropriately consented and/or assented. The subjects were compensated for their travel expenses. Inclusion criteria were a diagnosis of SMA Type 2, use of a power wheelchair for mobility, and between 5 and 19 years of age. Exclusion criteria were excessive UE joint contractures.

The participants' ages ranged from 8 to 19 years. At the start of the study, none of the subjects were on the Spinraza [nusinersen] medication trial (Biogen, 2020), but by the end of the study, two had started a trial of the new medication. One of the subjects who had started Spinraza participated in the study for the first 6 months and then withdrew. It is unclear if this was because of changes in medical or functional status resulting from the new medication or if the subject's withdrawal was unrelated. Spinraza is a relatively new pharmaceutical treatment with promising effects on slowing progression of disease and possibly restoring some muscle function (Biogen, 2020). Some improvement in hand function was seen at the end of this study in the remaining child trying Spinraza, but no difference was seen proximally. In future studies, this variable would have to be considered.

All five of the subjects chose to trial the WREX on both arms. Four of the subjects had the devices mounted to their wheelchair, and one had the devices mounted to an activity chair. The patients were fitted with custom bilateral WREX devices on their first visit (one was fitted with the second device a month later) (see Figure 2). They were educated on wear schedule and provided with activity ideas to get started. All of the subjects were asked to wear the WREX for at least 1 to 2 hr per day on average during the course of the study. Estimated daily wear times were reported at each visit or telephone call. The time of

day and setting were their choice. The parents were educated on how to don and doff the WREX and how to maintain proper alignment over time. All testing was performed in the research laboratory at our institution by the same occupational therapist. The following tests were performed at each time point: baseline, 6 months, and 1 year.

Figure 2

Participant in the Pilot Study Using Bilateral WREX Devices

Testing Instruments

Box and Block Test

The Box and Block Test (BBT) is a standardized, norm-referenced dexterity test widely used in occupational therapy with patients across the life span (Mathiowetz et al., 1985; Platz et al., 2005). The BBT counts how many blocks are moved from one side of the tray to the other over a barrier in 1 min. We administered the BBT to measure each subject's manual dexterity with dominant and non-dominant hands. The test was administered again with the WREX. Arm supports were removed from the wheelchair during this task to eliminate interference with the WREX. The BBT tray was placed on each subject's lap with support. Rest breaks were provided as needed between tasks to mitigate fatigue. Qualitative observations, such as need for propping on the edge of the box, wrist stability, and hand function were recorded for each subject. As the subjects' performance over time was the interest of this study, norms were not used.

Canadian Occupational Performance Measure (COPM)

The COPM is a widely used evidenced-based outcome measure designed to measure a patient's perception of his or her ability to engage in the skills of everyday living (Chan & Lee, 1997; Cup et al., 2003; Law et al., 1998). A semi-structured interview was conducted with each subject and his or her family. With this interview, we identified up to five meaningful activities in which the subject desired greater ability to participate. Self and family rating included current ability to perform these activities and associated level of satisfaction. A score of 1 reflected inability to perform that activity or high dissatisfaction with performing that activity. A score of 10 reflected the optimal performance of the task or the highest degree of satisfaction relating to performing that activity. For each subject, the scores for

performance on all five activities were averaged for each subject, and the scores for satisfaction on all five activities were averaged for each subject. The original activities chosen by each subject were reassessed at each visit. A change of two or more points on the average performance scale or satisfaction scale over time was considered clinically meaningful. Performance and satisfaction ratings were given for both with and without the WREX (first visit could only give ratings without use of the WREX).

Range of Motion

Shoulder range of motion (ROM) was measured with standard goniometry from a seated position in the subject's wheelchair. Each subject was assessed for presence of joint contractures of the UE at each visit (deficit of passive ROM) and current active range of motion (AROM) of left and right shoulders. In addition, a Microsoft Kinect (Microsoft, n.d.) sensor was used to motion-capture patients performing UE movements in all planes while seated in a chair. Each subject was asked to move their arms in a standardized sequence of movements using a video for them to follow. This sequence was repeated for both arms and again for both arms while wearing the WREX. Rest breaks were provided between each sequence. The motion-capture software was developed at the University of California, Davis (Han et al., 2013). The program was licensed to our team for this experiment. Movements were repeated for right and left arms both with and without the WREX. When the movements were detected by the Kinect, the software was used to calculate values describing movement in functional quadrants called reachable surface area (Han et al., 2015; Kurrillo et al., 2012; Oskarsson et al., 2016; Rammer et al., 2014). Each of these movement sequences was also video recorded in case of technical difficulties with the software.

Non-Standardized Questions Regarding Use

The families were asked at the final visit to comment on barriers to use, design issues, positive experiences, ability to participate in various settings, overall health status, and reports from other caregivers helping with use of the WREX. These questions were designed to reflect consideration for the World Health Organization International Classification of Functioning, Disability and Health scales with questions related to body components, activities and participation, and contextual factors (World Health Organization, 2001). The BBT, COPM, and ROM measurements were performed at each of the three visits.

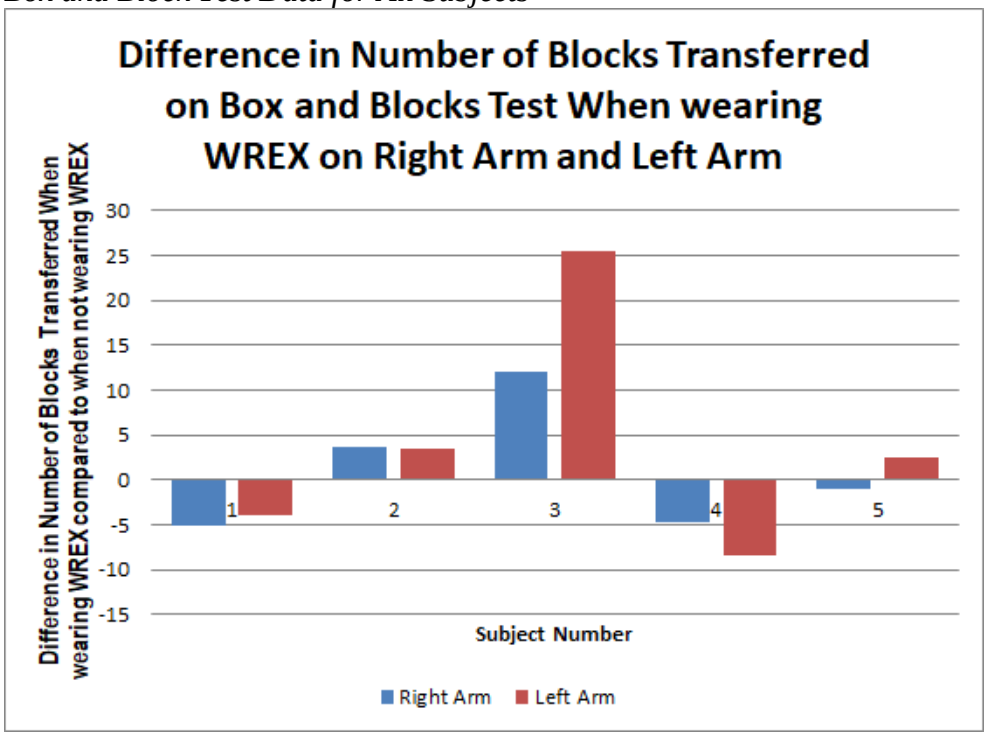
Results

Box and Block Test (BBT)

This test collected data for both hands, with and without the WREX. The number of blocks were averaged over two or three trials (two subjects did not complete the third trial [see Figure 3]). The data on the BBT performance revealed no clear indication that wearing the WREX consistently improves dexterity. For Subject 3, there was a large improvement in score when wearing the WREX bilaterally. However, data for Subjects 1 and 4 show how the WREX can interfere with performance. The standard deviations were large, and the data did not yield statistically significant differences between with and without the WREX.

Figure 3

Box and Block Test Data for All Subjects



Note. A positive bar indicates an improvement in dexterity with the WREX.

Canadian Occupational Performance Measure (COPM)

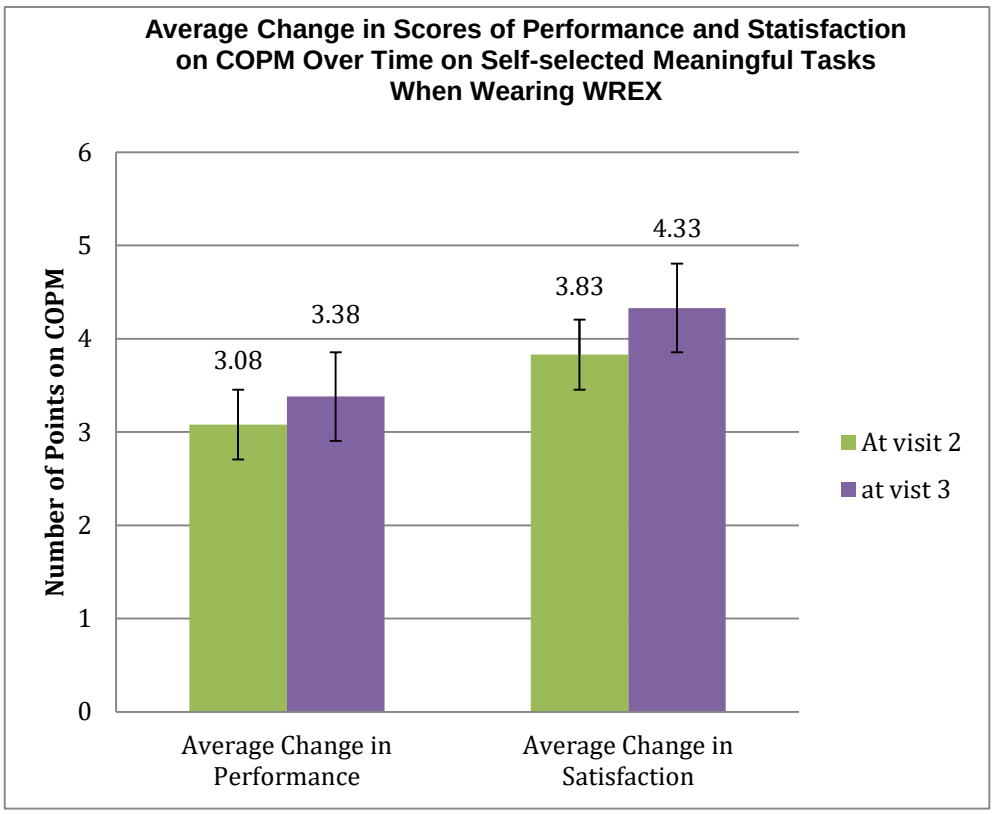
Each subject identified five occupations or activities that were meaningful to them and that they struggled to perform without a device, rating each with a score on performance and on satisfaction as described earlier. The performance scores were averaged for all tasks, and the satisfaction scores were also averaged separately. This was done for each subject at each visit rating the same activities. A difference was calculated in the mean performance score between Visits 1 and 2 and the satisfaction score between Visits 1 and 2. Differences were calculated again with the scores provided on Visits 2 and 3 for each subject. Then, the differences in performance and satisfaction were averaged and the standard deviation calculated. Self-ratings of performance and satisfaction without using a device were constant through the study except in the two cases where performance on all tasks dropped for Subjects 4 and 5 as a result of rapidly worsening scoliosis and the need for spinal surgery. Post-surgery self-ratings for performance and satisfaction were the same as Visit 1 for these subjects. Figure 4 shows the average difference between wearing the WREX to complete an activity versus completing the activity without the WREX on Visits 2 and 3.

Range of Motion (ROM)

Information on active shoulder flexion ROM is provided in Figure 5. We calculated averages for active shoulder flexion of all subjects at the start and end of the study. A separate average was calculated for the active motion of the right and left shoulder when wearing the WREX at the first and last visits for each subject.

Figure 4

Performance and Satisfaction Scores for the COPM

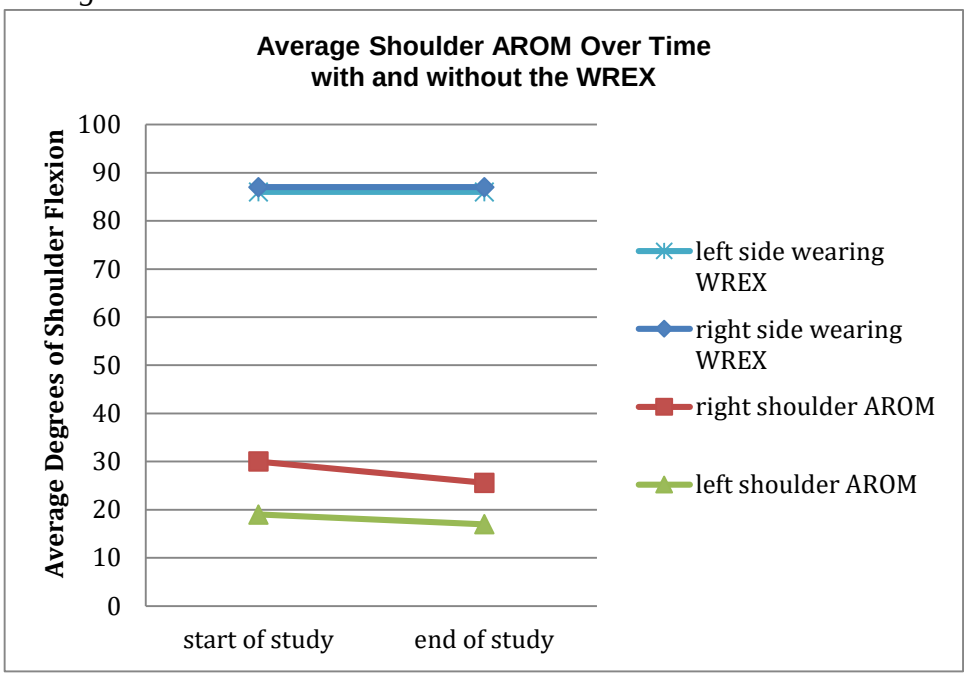


Note. Administered at each visit (clinically meaningful change established at 2 points).

Using standard goniometry, error in measurement is typically +/- 5 degrees. There was a slight decline in active motion over time across all subjects on the right side (16%) and left side (5%). Active ROM at the shoulder over time stayed constant when the participants were wearing the WREX. There was a slight decrease in performance of the shoulder when wearing the WREX just before the spinal fusions on two of the subjects. Increasing scoliosis prevented good alignment in these cases before surgery, but alignment was restored post surgery. Two of the subjects had contractures of less than 25 degrees in elbow flexion, and these contractures remained constant throughout the course of the study. The shoulder AROM measures with and without the WREX were significantly different; however, the AROM over the course of the study did not change.

Figure 5

Average Shoulder AROM Over Time with and without the WREX



Note. AROM = Active range of motion.

In addition, more subjective findings on posture for each subject are given as follows:

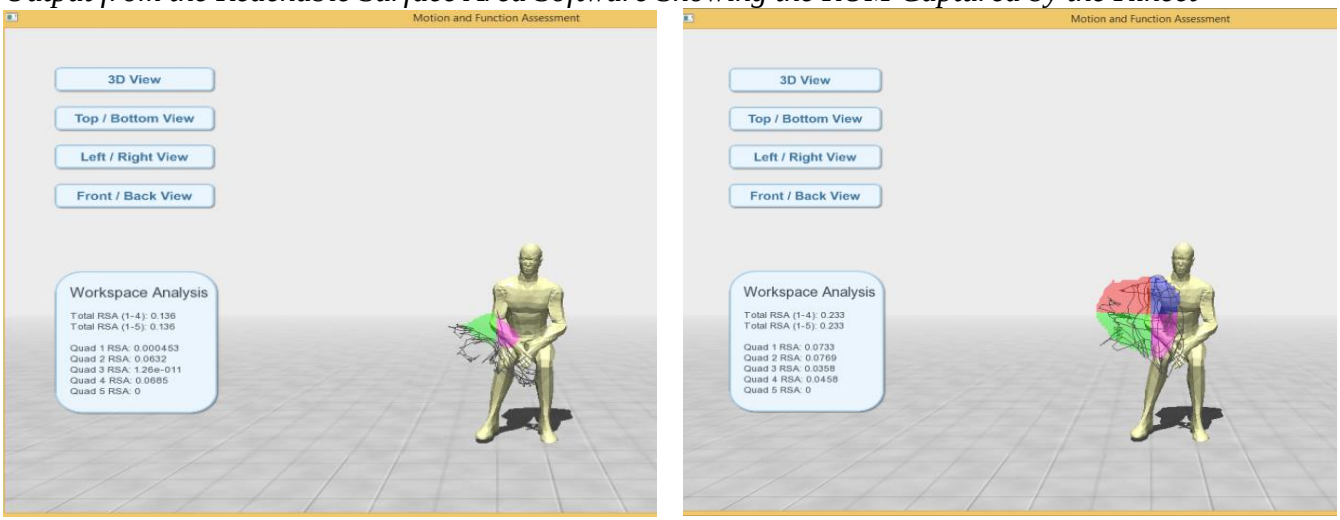
- Subject 1: Relatively strong postural control and good hand function. No upper extremity contractures were present.
- Subject 2: Poor postural control. Head often dropped down and she was unable to lift it up again. Poor hand function. Bilateral elbow flexion contracture of 10 degrees at the start and end of the study.
- Subject 3: Required a chest strap but had good head control and good hand function. Bilateral elbow flexion contractures of 25 degrees. No supination available on the right side. Mild wrist flexion contracture present at the start of the study. All contractures remained constant through study time period.
- Subject 4: Worsening postural control throughout study. She started wearing a scoliosis brace and then had spine surgery. Physical therapist noted increase in muscle activation after the WREX use was initiated. No contractures.
- Subject 5: Worsening trunk control was noted between visits at 6-month follow up. She started wearing a scoliosis brace then had spine surgery. No contractures were noted.

Reachable Surface Area

The Microsoft Kinect sensor was tried for each of the five subjects. For three of the subjects, the Kinect sensor generated no data. The data from the reachable workspace analysis software were essentially unusable because of inconsistencies in the Kinect sensor's ability to detect the children while sitting in their wheelchair. For one of the subjects, the Kinect did work well. Shown in Figure 6 is the workspace with and without the WREX.

Figure 6

Output from the Reachable Surface Area Software Showing the ROM Captured by the Kinect



Note. The ROM is broken up into quadrants. On the left is the subject without the WREX. To the right is the same subject with the WREX.

Discussion

This is the first study that tracked 1 year of use of the WREX in children with SMA. Many insights were gleaned from the objective findings and interviews. The paragraphs below address the research questions posed earlier.

Dexterity

There is a long-standing assumption that if there is better proximal stability, there will be greater distal control. Using the BBT, we theoretically gave the subjects better use of their shoulders to lift the blocks over the barrier to put on the targeted side via use of the WREX. All of the subjects were immediately able to do this without propping their arms on the edge of the box to use as a lever; all required propping of their arms when the UE was unassisted. Aside from this observation, no observable change in hand function or dexterity was noted when the shoulder was thus assisted. So, if the subject had poor hand function initially, the WREX improved quality of motion by positioning the hand as desired, but it did not impact actual dexterity. In one case, clinically meaningful gains in dominant hand use were made, but this was true with and without the WREX. This subject had started Spinraza medication. There were some data to suggest that improved motor learning may have occurred between Visits 1 and 2, which is when the WREX use was greatest in all of the subjects. It is unclear if the improvements would be maintained in the future if the WREX use was continued.

Function

Functionally speaking, the subjects all reported clinically meaningful improvements in performance of and satisfaction with the activities that they chose as important at the beginning of the study. The ability to participate in a greater variety of leisure interests was found to have the greatest improvement in both performance ratings (skill) and satisfaction ratings. The subjects were pleased to participate in music education (drumming), gym class, ball games, and crafts more often than they could previously. Facial hygiene or the ability to move a piece of hair away from the face or to scratch an itch on the face was also highly rated. Tasks such as writing and keyboard use had improved performance, but satisfaction was limited because of interference of the WREX with the table. The subjects reported being

able to tolerate wearing the WREX anywhere from 30 min to 2 hr at a time (limited by fatigue), but this time length decreased between the 6-month and 1-year visits because of a variety of medical complications.

Most of the families seem to assign one caregiver to use the WREX with the child in one setting over time. In some cases, the family identified self-feeding as a primary goal, but they did not attempt to use the WREX during family mealtimes, which was a little intriguing. This small study highlighted the incredible stresses and barriers a family with a child with SMA face over the course of a year and why multiple supports and therapy assistance may be needed to provide training on integrating WREX use into daily routines, learning how to use the arms in novel ways, and managing fatigue.

Range of Motion (ROM)

There were no obvious patterns of loss or gain of shoulder AROM over the course of the year. Variations in the AROM without a device can be explained primarily by worsening scoliosis over the course of the year leading two subjects to require spinal fusions during the last 6 months of this study. There was a clear improvement of shoulder AROM when wearing the WREX. Worsening kyphosis was noted to inhibit use of the WREX because of the inability to establish good alignment with the shoulder. Poor neck control and loss of ability to keep visual regard of hands affected functional use of the UEs both with and without the WREX. Use of a chest strap for postural support on a power chair did not impede use of the WREX. These postural changes were seen in the two youngest subjects, both of whom were 8 years of age. Both subjects noted with joint contractures at the elbow at the beginning of this study maintained baseline passive ROM throughout the study period. No subjects developed contractures during the study.

Data from the trials of reachable workspace analysis were not usable. While the Kinect sensor was able to detect and generate data from a wide selection of children and adults in trials prior to this study, it was unreliable in detecting children in power chairs during this study. It could detect movement in only two of the five subjects. We attempted to change the contrast of the background, to change the tilt of the chair, and to change the contrast of the child's clothing with the wheelchair. None of these changes yielded better results. Each sequence of movements was also observed and videotaped by the research team. When these videos were reviewed, we noted that in all subjects, the WREX allowed immediate functional movement patterns in the area from hip level to shoulder level with ease and, in some cases, to well above shoulder level. We also observed fewer compensatory movements of the trunk when the WREX were worn. A physical therapist working in the schools with one of the subjects did report that she noticed a significant improvement in muscle activation after use of the WREX began.

Overall, there is no evidence that during the course of 1 year the WREX had either a positive or a negative impact on ROM of the UE in this small population. Studies on the natural progression of SMA through childhood would be helpful to determine at what age it would be optimal to begin wearing a device such as the WREX. Would early use of the WREX promote better postural strength, delaying the need for spinal rods? Would use initiated before contractures develop prevent future contractures in the upper extremities? Would assisted movement with less compensation lead to less musculoskeletal pain (e.g., shoulder subluxations and cervical pain) in adulthood? These questions remain unanswered.

The families were asked a variety of questions at the end of the study relating to International Classification of Functioning categories of body function and structure, activities and participation, and environmental factors. Four of the five families were able to answer these questions and participated in a

dialogue about the experience over 1 year (one subject dropped out before the final session). Common themes in questions related to body function and structure were as follows:

- The overall health of these five subjects varied, but frequent hospitalizations and fatigue were common factors that affected their ability to wear the device and participate in timely follow-up visits. Four subjects who completed exit interviews had at least one surgery during the course of the year-long study. All of the subjects also had at least one, and up to four, hospitalizations not related to their surgeries.
- Two of the subjects reported that they would choose the bilateral WREX devices again, and all wanted to keep the devices after the study. One said she preferred to use the WREX on the nondominant side because the dominant-side WREX interfered with driving the powered WC. One said she preferred to use the dominant side only because using both at the same time felt cumbersome.
- The subjects reported fatigue when wearing the device from 30 min to 120 min (average was 78 min).
- One of four subjects reported wearing a wrist brace for support or use of an assistive device in the hand like a universal cuff to support hand function.
- The subjects rated their ability to don and doff the device (1 = *very difficult*, 5 = *very easy*). Average score was 4.75.
- The subjects rated their image of self when wearing the device(s) (1 = *I am embarrassed to wear it*, 5 = *I really liked the way I looked*). Average score was 3.5.

Common themes in the questions related to activity and participation were as follows:

- Four of the subjects reported being able to reach their hand to desired targets in the environment.
- All reported the WREX increased their participation in personal activities. They identified personal activities as hygiene-related, meal preparation, dressing self as a princess, and moving obstacles in the environment as needed.
- All reported WREX use increased their participation in social activities (e.g., ball games, indoor recess games, board games with friends, and being able to eat popcorn in the movie theatre for the first time).
- Two had therapy during the course of this study, but the WREX was used during therapy in only one case.
- Two reported improvements in participation in school, citing increased ability to raise their hand and be recognized and also improvement in gym, music, art, and recess.
- All reported improvements in participation in the home environment (e.g., use of iPad, play board games with siblings, meal preparation, eating, and crafts).
- When asked what their friends thought of the device, all reported their friends thought it was cool or that they liked that it allowed them to do an activity together.

Common themes in the questions related to environmental factors were as follows:

- The subjects were asked where the WREX was primarily used. Two reported school and home, one reported home only, one reported home and community.
- The subjects were asked about physical barriers to use. They reported the WREX hit tables when doing desk work, hit doorways, and interfered with armrest on WC and driving joystick.

- The subjects were asked if it was difficult to maintain alignment of the devices. All said no, except if there was a need to change to a different wheelchair. One said it took a little practice, but it was not difficult.
- The subjects were asked if they trained other caregivers to use the WREX. Two said their mothers were the only caregivers using the WREX, one said home nurses were trained, and one had a 1:1 school aide trained.

Study Limitations

This study has several limitations. The sample size is too small to generalize or demonstrate statistical significance. The data support the trends in function identified in other studies (Gunn et al., 2016; Shank et al., 2017) and provide new insight into the challenges of long-term use. Future studies should recruit a larger cohort of subjects so that generalizations can be made. In addition, there are no validation studies for use of the BBT with this specific population, and this is not likely to happen in this small a population. So, we relied on the expert opinions of two certified hand therapists working in pediatrics and a pediatric hand surgeon in our decision to use this measure. Range of motion data, specifically reachable surface area data, were unattainable because of technical difficulties. In discussions with the parents about wear time, subjective parent/subject report had to be used. In future studies, a daily diary or compliance sensor would be more accurate. Personal factors were also out of our control during the year-long study. Initiating medications in the middle of this study, as well as surgeries, was not ideal for controlled conditions; however, the study does show what can happen during a given year for medically complex children and so gives insight into the challenges and barriers these children face.

Conclusion

From the experience of these five children who attempted to wear the WREX for about 1 hr per day for 1 year, we offer a few conclusions. Wearing the WREX devices did dramatically increase the active ROM at the shoulder immediately. There did not appear to be improvement in AROM while wearing the WREX over time. Wearing the WREX over the course of 1 year did not result in development of any contractures, nor did it improve existing contractures. The children wearing the WREX when completing the BBT exhibited fewer postural compensations (e.g., leaning) and distal compensations (e.g., propping the arm). However, this did not consistently lead to better ability to grasp and release the blocks (no consistent improvement in score). All of the subjects reported gains in functional performance and satisfaction in the activities they chose as meaningful, and these gains lasted or improved over the year. The subjects rated the largest improvements in performance and satisfaction for participation in leisure skills, followed by facial hygiene tasks and self-feeding tasks. All of the subjects asked to keep the WREX after the study was completed. Common barriers to use were fatigue; difficulty in transport; worsening scoliosis; and illness, hospitalizations, and surgeries. Future studies should consider age and disease progression as factors in use of the WREX. Parents should be educated that during periods of decline (e.g., posture) the WREX may be contraindicated, but it will be useful again after spinal fusion. Discomfort and frustration can be avoided if expectations are established early that fatigue will likely occur after 30 min and appropriate rest breaks are necessary. Occupational therapy may be helpful to increase activity tolerance and endurance with the devices as well as to help caregivers overcome barriers in various environments.

References

Armon Products B. V. (1999). The armon [Equipment]. www.armonproducts.com

Arnold, W. D., Kassar, D., & Kissel, J. T. (2015). Spinal muscular atrophy: Diagnosis and management in a new therapeutic era. *Muscle & Nerve*, *51*(2), 157–167. https://doi.org/10.1002/mus.24497

Assistive Innovations. (n.d.). The dynamic arm support [Equipment]. https://www.assistive-innovations.com/en/arm-supports

Biogen. (2020). Spinraza [nusinersen]. Cambridge, MA. https://www.spinraza.com/

Chan, C. C. H., & Lee, T. M. C. (1997). Validity of the Canadian occupational performance measure. *Occupational Therapy International*, *4*(3), 145–166. https://doi.org/10.1002/oti.58

Cup, E. H., Scholte op Reimer, W. J., Thijssen, M. C., & vanKuyk-Minis, M. (2003). Reliability and validity of the Canadian occupational performance measure in stroke patients. *Clinical Rehabilitation*, *17*(4), 402–409. https://doi.org/10.1191/0269215503cr635oa

Gunn, M., Shank, T. M., Eppes, M., Hossain, J., & Rahman, T. (2016). User evaluation of a dynamic arm orthosis for people with neuromuscular disorders. *IEEE Transactions in Neural Systems and Rehabilitation Engineering*, *24*(12), 1277–1283. https://doi.org/10.1109/tnsre.2015.2492860

Han, J. J., Kurillo, G., Abresch, R. T., Nicorici, A., & Bajcsy, R. (2013). Validity, reliability and sensitivity of a 3D vision sensor-based upper extremity reachable workspace evaluation in neuromuscular diseases. *PLOS Current Dec*, *12*(5). https://doi.org/10.1371/currents.md.f63ae7dde63caa718fa0770217c5a0e6

Han, J. J., de Bei, E., Nicorici, A., Abresch, R. T., Bajcsy, R., & Kurillo, G. (2015). Reachable workspace reflects dynamometer-measured upper extremity strength in facioscapulohumeral muscular dystrophy. *Muscle & Nerve*, *52*(6), 948–955. https://doi.org/10.1002/mus.24651

Haumont, T., Rahman, T., Sample, W., King, M., Church, C., Henley, J., & Jayakumar, S. (2011). Wilmington robotic exoskeleton: A novel device to maintain arm improvement in muscular disease. *Journal of Pediatric Orthopeadics*, *31*(5), e44–e49. https://doi.org/10.1097/bpo.0b013e31821f50b5

Herder, J. L., Vrijlandt, N., Antonides, T., Cloosterman, M., & Mastenbroek, P. L. (2006). Principle and design of a mobile arm support for people with muscular weakness. *Journal of Rehabilitation Research and Development*, *43*(5), 591–604. https://doi.org/10.1682/jrrd.2006.05.0044

JAECO Orthopedic. (n.d.). JAECO Wilmington Robotic Exoskeleton. https://jaecoorthopedic.com/product/jaeco-wrex/

Kurrillo, G., Han, J. J., Abresch, R. T., Nicorici, A., Yan, P., & Bajcsy, R. (2012). Development and application of stereo camera based upper extremity workspace evaluation in patients with neuromuscular diseases. *PLOS One*, *7*(9), e45341. https://doi.org/10.1371/journal.pone.0045341

Law, M., Baptiste, S., Carswell, A., McColl, M., Polatajko, H., & Pollock, N. (1998). *Canadian Occupational Performance Measure* (Rev. 2nd ed.). Canadian Association of Occupational Therapists Publications ACE.

Mathiowetz, V., Ferderman, S. M., & Wiemer, D. M. (1985). Box and block test of manual dexterity: Norms for 1–19 year-olds. *Canadian Journal of Occupational Therapy*, *52*(5), 241–246. https://doi.org/10.1177/000841748505200505

Microsoft. (n.d.). Kinect for windows. https://developer.microsoft.com/en-us/windows/kinect/

Oskarsson, B., Joyce, N. C., De Bie, E., Nicorici, A., Bajcsy, R., Kurillo, G., & Han, J. J. (2016). Upper extremity 3-dimensional reachable workspace assessment in amyotrophic lateral sclerosis by Kinect sensor. *Muscle & Nerve*, *53*(2), 234–241. https://doi.org/10.1002/mus.24703

Platz, T., Pinkowski, C., van Wijck, F., Kim, I. H., di Bella, P., & Johnson, G. (2005). Reliability and validity of arm function assessment with standardized guidelines for the Fugl Meyer test, action research arm test, and box and block test: A multicenter study. *Clinical Rehabilitation*, *19*(4), 404–411. https://doi.org/10.1191/0269215505cr832oa

Rahman, T., Sample, W., Jayakumar, S., King, M. M., Wee, J. Y., Seliktar, R., Alexander, M., Scavina, M., & Clark, A. (2006). Passive exoskeletons for assisting limb movement. *Journal of Rehabilitation Research Development*, *43*(5), 583–590. https://doi.org/10.1682/jrrd.2005.04.0070

Rahman, T., Sample, W., Seliktar, R., Scavina, M. T., Clark, A. L., Moran, K., & Alexander, M. A. (2007). Design and testing of a functional arm orthosis in patients with neuromuscular diseases. *IEEE transactions on Neural Systems and Rehabilitation Engineering*, *15*(2), 244–251. https://doi.org/10.1109/tnsre.2007.897026

Rammer, J. R., Kzrak, J., Riedel, S. A., & Harris, G. F. (2014). Evaluation of upper extremity movement characteristics during standardized pediatric functional assessment with a Kinect-based markerless motion analysis system. *2014 36th Annual International Conference of the IEEE Engineering in Medicine and Biology Society*, https://doi.org/10.1109/embc.2014.6944136

Saebo Inc. (n.d.). SaeboMAS. https://www.saebo.com/shop/saebomas/

Shank, T., Eppes, M., Hossain, J., Gunn, M., & Rahman, T. (2017). Outcome measures with COPM of children using a Wilmington robotic exoskeleton. *The Open Journal of Occupational Therapy*, *5*(1). https://doi.org/10.15453/2168-6408.1262

Spinal Muscular Atrophy. (2019, January 28). *Genetic and rare diseases information center (GARD)*. http://rarediseases.info.nih.gov/gard/7883/spinal-muscular-atrophy-1/resources/1

Van der Heide, L. A., Van Ninhuijs, B., Bergsma, A., Gelderblom, G. J., van der Pijl, D. J., & de Witte, L. P. (2014). An overview and categorization of dynamic arm supports for people with decreased arm function. *Prosthetics and Orthotics International*, *38*(4), 287–302. https://doi.org/10.1177/0309364613498538

World Health Organization. (2001). *International classification of functioning, disability and health: ICF*. https://apps.who.int/iris/handle/10665/42407